B. B. Sharma¹, Shashi Sharma², Naveen Bhardwaj³, Pankaj Abrol⁴, Sakshi Dewan⁵, Mir Rizwan Aziz⁶
¹Professor & HOD, Dept. of Radio diagnosis, SGT Medical College Budhera (Gurgaon).
²Associate Professor, Department of Pediatrics, SGT Medical College, Gurgaon (Haryana)
³Assistant Professor, Dept. of Radio diagnosis, SGT Medical College Budhera (Gurgaon).
⁴Professor & HOD, Dept.of Pediatrics, SGT Medical College Budhera (Gurgaon).
⁵Senior Resident, PGIMER & RML Hospital,New Delhi -110001.
⁶Senior Resident, Dept. of Radio diagnosis, SGT Medical College Budhera (Gurgaon)
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ABSTRACT
There is wide spectrum of Mullerian dysgenesis presenting in different ways. Routine ultrasound done of a child for pain abdomen discovered a pelvic mass supero-posterior to the urinary bladder. This was diagnosed as fused pelvic kidneys by various cross sectional imaging modalities like Computerized tomography (CT) and Magnetic resonance imaging (MRI). We present a 4-years male child who was diagnosed as a case of “lump kidneys” by plain sonography coupled with color flow imaging (CFI). The entity usually falls in the common group of VATER (vertebral, anorectal malformation, esophageal and renal) anomalies. But our present case was having isolated anomaly and this entity is of a great rarity as seen in literature.

Keywords: Mullerian dysgenesis, Ultrasound, lump kidneys, CFI, CT, MRI, VATER.

INTRODUCTION
The pelvic kidney is also called as sacral kidney because of the location. This ectopia is seen in 1 out of 2100-3000 autopsy cases. These cases may present with atypical features. The entity remains asymptomatic for a long time until the diagnosis is made by imaging modalities after some vague complaints.

CASE REPORT
4-years old male child [Figure 1] was brought to the children outpatient department by the parents with the complaint of pain lower abdomen of one month duration.
The child was full term first sibling and was born by normal delivery. There was no history of any maternal health problem before or during pregnancy. On examination child weighed 10 kg and was of normal physical and growth parameters. On palpation of abdomen there was firm non mobile lump in the upper part of pelvic region having mild tenderness. Rest of the systemic examination was unremarkable. Blood urea and serum creatinine were 20 mg% and 0.64 mg% respectively. Routine urine examination did not show any infection. All biochemical parameters were within normal limits. Plain X-ray abdomen and pelvis were normal [Figure 2].

Figure 1: Photograph of 4-years old male child with normal physique and outlook in respect of height, weight and physical development. No visible physical abnormality was noticed in any of the four limbs.
Ultrasound examination has shown empty bilateral renal fossae. There was presence of pelvic mass postero-superior to the urinary bladder. Color flow imaging (CFI) revealed the vascular structures within this pelvic mass as that of kidneys. Both the renal arteries were taking origin from the common iliac arteries of the respective sides [Figure 3a,b & 4a,b].

Though the intravenous pyelography was the investigation of choice for the further evaluation but CT urography was done to rule out the group of VATER anomalies. The proper prior consent of the parents was taken before the investigations. CT urography revealed bilateral normal functioning fused kidneys in the pelvis region. These kidneys were draining by their separate small ureters without any congenital abnormality [Figure 5a and b].

This was further delineated as fused pelvic kidneys by MR Urography. Both the fused kidneys have pelvis pointing to the supero anterior axis and ureters draining to the bladder by their short course. The bladder was well delineated without any diverticulum or any other abnormality. There was no dilatation of either of the ureters. Bladder neck was also well depicted without any extrinsic impression or intrinsic filling defect [Figure 6a, b and c].

**DISCUSSION**

Pelvic fused kidney is a developmental anomaly and is of rare type in nature. It is very important to understand the embryogenesis of the developing kidney to know in depth regarding this anomaly. Both the kidneys develop from the nephron blastema present in the pelvis at the level of sacral somites. The various factors which play in the further
migration, permanent positioning and functioning can be disturbed leading to various anomalies and "pelvic fused kidneys" is one among those varieties. Three factors which play the role in proper development and positioning are as follows:

- Ascent
- Rotation
- Vascular supply

Umblical arteries can obstruct the ongoing ascent of the kidneys to the umbilical region. By 8th week both the kidneys should be placed at their normal lumbar region location. Various anomalies can arise because of disturbance of these factors. The diagrammatic representation of the layout of the fused or pancake kidney can be explained and compared with their normal layout in relation to the major vessels in the pelvis region [Figure 7a and b].

Figure 5: Contrast Enhanced Computerized Tomography of the pelvic region. (a) upper region of the fused kidneys show anteriorly projecting pelvis of both the kidneys (vertical upward black arrow). (b) lower section shows opacified pelvicalyceal system with prominent upper part of the left ureter (white downward pointing arrow).

Figure 6: MR urography. (a) coronal anterior section shows right ureter (thin white arrow) and slightly prominent left ureter (thick wide arrow) coursing towards the urinary bladder. (b) coronal section at slightly posterior aspect shows separate masses of both the kidneys. (c) sagittal section shows right uretero-vasical junction (wide black arrow) and left uretero-vasical junction (horizontal white arrow).

There can be various permutations and combinations in the form of ectopia or fused kidneys and that too at different locations. The initial imaging modality is the ultrasound study coupled with color Doppler studies. This outlines the anatomical layout of the kidneys and their vasculature respectively. Functional status of the fused kidney can be verified with the help of intravenous pyelography or computerized tomography urography. Magnetic resonance imaging can be helpful in outlining other organs in relation to kidneys. The modality is free of any radiation and urography can be done without any administration of contrast.

The anomaly remains asymptomatic for a long time until some complication develops either because of the anomaly itself or infection and stone formation. The incidence of reflux and stone formation is very high with the background of underlying anomalies. The surgical maneuver can be contemplated through laproscopic procedure if any complication is there.
CONCLUSION

Pelvic fused kidneys may not be diagnosed early because of no complaints. This may present as vague abdominal pain and routine ultrasound examination shows empty renal fossae and presence of the mass in the pelvic region. Radiological evaluation by US, CFI, CT Urography and MR Urography can clearly guide the clinician for further management. It is imperative to rule out other VATER anomalies related to this anomaly.

REFERENCES


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